SICKLE-CELL ANEMIA AND SICKLE-CELL TRAIT

BASIC INFORMATION

DESCRIPTION

An inherited blood disorder that causes anemia, episodes of severe pain, low resistance to infection and chronic poor health. It is not cancerous. It involves the bone marrow, lymph glands, spleen, liver, and thymus, usually begins around 6 months of age and lasts lifelong.

FREQUENT SIGNS AND SYMPTOMS

- Anemia with shortness of breath, rapid heartbeat, fatigue and jaundice.
- Episodes of pain in joints, chest, abdomen and back.
- Frequent infections, especially pneumonia.
- Nerve impairment.
- Delayed growth and development.
- Skin ulcers, especially on the legs.

CAUSES

This disease is hereditary. The red blood cells contain an abnormal type of hemoglobin called hemoglobin S. Persons with the hemoglobin S may pass it on to their children. Red blood cells change from round to sickle shapes, which causes blockage in the capillaries. Low oxygen in the tissues is partly responsible for the changed shape. The change occurs in attacks that cause pain and disability ("sickle crisis"). The disease occurs mostly in black people.

RISK INCREASES WITH

Family history of sickle-cell anemia. The following may aggravate symptoms:

- Ascending to high altitude, as in driving up a mountain or flying.
- Pregnancy.
- Surgery.
- Injury.
- Infection.

PREVENTIVE MEASURES

- If you have a family history of sickle-cell anemia, ask for testing. If the condition is present, obtain genetic counseling before starting a family. A less serious condition, sickle-cell trait, may be present. It will not cause the disease, but genetic counseling is still desirable.
- Tests in early pregnancy to determine if unborn child has inherited the double-dose gene (both parents are carriers).

EXPECTED OUTCOMES

Sickle-cell anemia is incurable and life expectancy is reduced. However, life span has gradually increased to over 40 years with increasingly effective treatments. Most patients die prematurely of infection or stroke.

POSSIBLE COMPLICATIONS

- Persons with sickle-cell trait may be at risk of sudden death while engaged in strenuous exercise.
- Infections of lungs and bones.
- Kidney failure.
- Eye disease.
- Stroke.

TREATMENT

GENERAL MEASURES

- Diagnostic tests may include laboratory blood studies, X-rays, MRI or CT scan of bones and lungs. Simple screening tests are also available. They may be done at birth if there is a family history of sickle-cell anemia.
- If a child has the condition, a referral to a doctor with special knowledge of this condition may be recommended.
- Treatment at home involves general health care maintenance and prompt treatment of sickle cell crises.
- Hospitalization may be required at times of severe attacks for intravenous therapy and oxygen therapy and sometimes, blood transfusions.
- During an attack, help patient stay warm. Apply warm compresses to painful areas.
- Maintain immunization schedule, including a pneumonia vaccine.
- Don't fly, even in pressurized planes, without oxygen. Check with your airline.
- Wear a medical alert type bracelet or pendant to identify the medical disorder.
- Psychotherapy or counseling may be helpful in adapting to this condition, especially for children.
- Additional information available from the National Association for Sickle Cell Disease, 3345 Wilshire Blvd., Suite 1106, Los Angeles, CA 90010-1880, (800) 421-8453.

MEDICATIONS

No medications are yet available to control this condition. For severe attacks, intravenous fluids, blood transfusions, antibiotics and pain relievers may be used. Prophylactic penicillin may be started in infancy. For recurrent painful attacks, hydroxyurea has been shown to reduce the frequency of episodes.

ACTIVITY

- Avoid strenuous exercise and exposure to cold temperatures. Rest in bed during acute attacks.
- Activity may be somewhat limited due to chronic anemia and poor muscular development.

DIET

Drink at least 8 glasses of water a day or more if there is a fever. This helps keep blood cells from collecting and blocking capillaries.

NOTIFY OUR OFFICE IF

- Your child has signs and symptoms of sickle-cell anemia.
- You want to know if you have the sickle-cell gene.
- You have the disease, and symptoms recur after a period of remission or you develop fever or infection.